



## General

### Guideline Title

Motor neurone disease: assessment and management.

### Bibliographic Source(s)

National Clinical Guideline Centre. Motor neurone disease: assessment and management. London (UK): National Institute for Health and Care Excellence (NICE); 2016 Feb 24. 47 p. (NICE guideline; no. 42).

### Guideline Status

This is the current release of the guideline.

This guideline updates a previous version: Centre for Clinical Practice. Motor neurone disease. The use of non-invasive ventilation in the management of motor neurone disease. London (UK): National Institute for Health and Clinical Excellence (NICE); 2010 Jul. 127 p. (Clinical guideline; no. 105). [53 references]

This guideline meets NGC's 2013 (revised) inclusion criteria.

## Regulatory Alert

### FDA Warning/Regulatory Alert

Note from the National Guideline Clearinghouse: This guideline references a drug(s) for which important revised regulatory and/or warning information has been released.

- [March 22, 2016 – Opioid pain medicines](#) : The U.S. Food and Drug Administration (FDA) is warning about several safety issues with the entire class of opioid pain medicines. These safety risks are potentially harmful interactions with numerous other medications, problems with the adrenal glands, and decreased sex hormone levels. They are requiring changes to the labels of all opioid drugs to warn about these risks.

## Recommendations

### Major Recommendations

Note from the National Guideline Clearinghouse (NGC): This guideline was developed by the National Clinical Guideline Centre (NCGC) on

behalf of the National Institute for Health and Care Excellence (NICE). See the "Availability of Companion Documents" field for the full version of this guidance and related appendices.

The wording used in the recommendations in this guideline (for example, words such as 'offer' and 'consider') denotes the certainty with which the recommendation is made (the strength of the recommendation) and is defined at the end of the "Major Recommendations" field.

Recommendations are marked as:

- [new 2016] if the evidence has been reviewed and the recommendation has been added or updated
- [2010] if the evidence has not been reviewed since the original guideline
- [2010, amended 2015] if the evidence has not been reviewed but changes have been made to the recommendation wording that change the meaning (for example, because of equalities duties or a change in the availability of medicines, or incorporated guidance has been updated)
- [2010, amended 2016] if the evidence has not been reviewed but changes have been made to the recommendation wording that change the meaning

### Recognition and Referral

Ensure that robust protocols and pathways are in place to:

- Inform healthcare professionals about motor neurone disease (MND) and how it may present
- Inform healthcare professionals in all settings about local referral arrangements
- Ensure continued and integrated care for people with MND across all care settings [new 2016]

Be aware that MND causes progressive muscular weakness that may first present as isolated and unexplained symptoms. These symptoms may include:

- Functional effects of muscle weakness, such as loss of dexterity, falls or trips
- Speech or swallowing problems, or tongue fasciculations (this is known as bulbar presentation)
- Muscle problems, such as weakness, wasting, twitching, cramps and stiffness
- Breathing problems, such as shortness of breath on exertion or respiratory symptoms that are hard to explain
- Effects of reduced respiratory function, such as excessive daytime sleepiness, fatigue, early morning headache or shortness of breath when lying down [new 2016]

Be aware that MND may first present with cognitive features, which may include:

- Behavioural changes
- Emotional lability (not related to dementia)
- Frontotemporal dementia [new 2016]

If you suspect MND, refer the person without delay and specify the possible diagnosis in the referral letter. Contact the consultant neurologist directly if you think the person needs to be seen urgently. [new 2016]

Provide information and support for people and their family members and/or carers (as appropriate) throughout the diagnostic process, particularly during periods of diagnostic uncertainty or delay. [new 2016]

### Information and Support at Diagnosis

Please also refer to the recommendations in NICE's guideline on [patient experience in adult NHS services](#) , which includes recommendations on communication, information and coordination of care.

Information about the diagnosis, prognosis and management of MND should be given by a consultant neurologist with up-to-date knowledge and experience of treating people with MND unless it is clinically necessary to give the diagnosis in an urgent situation. The neurologist should have knowledge and expertise in the following:

- Symptoms of MND
- Types and possible causes of MND
- Treatment options
- How MND may progress (including cognitive and behavioural changes) and how progression may affect the treatments offered
- Crisis prevention (for example, if there is an acute hospital admission or a breakdown in care arrangements)
- Opportunities for people with MND to be involved in research

- Likely needs and concerns of people with MND and their family members and/or carers (as appropriate)
- Advance care planning [new 2016]

Ask people about how much information they wish to receive about MND, and about their preferences for involving their family members and/or carers (as appropriate). [new 2016]

Ensure people are provided with information about MND and support at diagnosis or when they ask for it. If the person agrees, share the information with their family members and/or carers (as appropriate). Information should be oral and written, and may include the following:

- What MND is
- Types and possible causes
- Likely symptoms and how they can be managed
- How MND may progress
- Treatment options
- Where the person's appointments will take place
- Which healthcare professionals and social care practitioners will undertake the person's care
- Expected waiting times for consultations, investigations and treatments
- Local services (including social care and specialist palliative care services) and how to get in touch with them
- Local support groups, online forums and national charities, and how to get in touch with them
- Legal rights, including social care support, employment rights and benefits
- Requirements for disclosure, such as notifying the Driver and Vehicle Licensing Agency (DVLA)
- Opportunities for advance care planning [new 2016]

When MND is diagnosed, provide people with a single point of contact for the specialist MND multidisciplinary team (see "Organisation of Care" section below). Provide information: about what to do if there are any concerns between assessments or appointments, during 'out-of-hours' or in an emergency, or if there is a problem with equipment. [new 2016]

Offer the person with MND a face-to-face, follow-up appointment with a healthcare professional from the multidisciplinary team, to take place within 4 weeks of diagnosis. [new 2016]

When MND is suspected or confirmed, inform the person's general practitioner (GP) without delay and provide information about the likely prognosis. [new 2016]

Set aside enough time to discuss the person's concerns and questions, which may include the following:

- What will happen to me?
- Are there any treatments available?
- Is there a cure?
- How long will I live?
- What will the impact on my day-to-day life be?
- What will happen next with my healthcare?
- Will my children get MND?
- How do I tell my family and friends?
- How will I die? [new 2016]

If the person has any social care needs, refer them to social services for an assessment. Be aware that some people with MND may not have informal care available, and may live alone or care for someone else. [new 2016]

Advise carers that they have a legal right to have a Carer's Assessment of their needs; support them with requesting this from their local authority. [new 2016]

### Cognitive Assessments

Please also refer to the recommendations in NICE's guideline on [patient experience in adult NHS services](#) .

Be aware that people with MND and frontotemporal dementia may lack mental capacity. Care should be provided in line with the Mental Capacity Act 2005. [new 2016]

At diagnosis, and if there is concern about cognition and behaviour, explore any cognitive or behavioural changes with the person and their family

members and/or carers as appropriate. If needed, refer the person for a formal assessment in line with the NICE guideline on [dementia](#) [new 2016]

Tailor all discussions to the person's needs, taking into account their communication ability, cognitive status and mental capacity. [new 2016]

### Prognostic Factors

When planning care take into account the following prognostic factors, which are associated with shorter survival if they are present at diagnosis:

- Speech and swallowing problems (bulbar presentation)
- Weight loss
- Poor respiratory function
- Older age
- Lower Amyotrophic Lateral Sclerosis Functional Rating Scale (ALSFRS or ALSFRS-R) score
- Shorter time from first developing symptoms to time of diagnosis [new 2016]

### Organisation of Care

Provide coordinated care for people with MND, using a clinic-based, specialist MND multidisciplinary team approach. The clinic may be community or hospital based. [new 2016]

The multidisciplinary team should:

- Include healthcare professionals and social care practitioners with expertise in MND, and staff who see people in their home
- Ensure effective communication and coordination between all healthcare professionals and social care practitioners involved in the person's care and their family members and/or carers (as appropriate)
- Carry out regular, coordinated assessments at the multidisciplinary team clinic (usually every 2 to 3 months) to assess people's symptoms and needs
- Provide coordinated care for people who cannot attend the clinic, according to the person's needs [new 2016]

The multidisciplinary team should assess, manage and review the following areas, including the person's response to treatment:

- Weight, diet, nutritional intake and fluid intake, feeding and swallowing (see "Nutrition and Gastrostomy" below)
- Muscle problems, such as weakness, stiffness and cramps (see recommendations under "Managing Symptoms" below)
- Physical function, including mobility and activities of daily living (see "Equipment and Adaptation to Aid Activities of Daily Living and Mobility" below)
- Saliva problems, such as drooling of saliva (sialorrhoea) and thick, tenacious saliva (see recommendations under "Managing Symptoms" below)
- Speech and communication (see "Communication" below)
- Cough effectiveness (see "Cough Effectiveness" below)
- Respiratory function, respiratory symptoms and non-invasive ventilation (see "Respiratory Function and Respiratory Symptoms and "Non-invasive Ventilation" below)
- Pain and other symptoms, such as constipation
- Cognition and behaviour (see "Cognitive Assessments" above)
- Psychological support needs (see recommendations under "Psychological and Social Care Support" below)
- Social care needs (see recommendations under "Psychological and Social Care Support" below)
- End of life care needs (see "Planning for End of Life" below)
- Information and support needs for the person and their family members and/or carers (as appropriate) (see "Information and Support at Diagnosis" above) [new 2016]

The core multidisciplinary team should consist of healthcare professionals and other professionals with expertise in MND, and should include the following:

- Neurologist
- Specialist nurse
- Dietitian
- Physiotherapist
- Occupational therapist

- Respiratory physiologist or a healthcare professional who can assess respiratory function
- Speech and language therapist
- A healthcare professional with expertise in palliative care (MND palliative care expertise may be provided by the neurologist or nurse in the multidisciplinary team, or by a specialist palliative care professional) [new 2016]

The multidisciplinary team should have established relationships with, and prompt access to, the following:

- Clinical psychology and neuropsychology
- Social care
- Counselling
- Respiratory ventilation services
- Specialist palliative care
- Gastroenterology
- Orthotics
- Wheelchair services
- Assistive technology services
- Alternative and augmentative communication (AAC) services
- Community neurological care teams [new 2016]

Tailor the frequency of the multidisciplinary team assessments to the person's symptoms and needs, with more or less frequent assessments as needed. [new 2016]

Ensure arrangements are in place to trigger an earlier multidisciplinary team assessment if there is a significant change in symptoms identified by the person, family members and/or carers (as appropriate), or healthcare professionals. [new 2016]

Tailor the multidisciplinary team assessment to the person's needs, for example, adjust the format if the person has cognitive or behaviour changes or difficulties with communication. [new 2016]

Inform all healthcare professionals and social care practitioners involved in the person's care about key decisions reached with the person and their family members and/or carers (as appropriate). [new 2016]

Ensure that all healthcare professionals and social care practitioners involved in the person's care are aware that MND symptoms may get worse quickly, and that people with MND will need repeated, ongoing assessments. Priority should be given to ensuring continuity of care and avoiding untimely case closure. [new 2016]

Consider referral to a specialist palliative care team for people with current or anticipated significant or complex needs, for example, psychological or social distress, troublesome or rapidly progressing symptoms and complex future care planning needs. [new 2016]

For guidance on the use of riluzole for people with MND, see the NICE technology appraisal [guidance on the use of riluzole \(Rilutek\) for the treatment of motor neurone disease](#) . [new 2016]

### Psychological and Social Care Support

During multidisciplinary team assessments and other appointments, discuss the psychological and emotional impact of MND with the person and ask whether they have any psychological or support care needs. Topics to discuss may include the following:

- Their understanding of MND and how it affects daily living
- Accepting and coping with the diagnosis and prognosis, including concerns and fears about dying
- Their ability to continue with current work and usual activities
- Adjusting to changes in their life and their perception of self
- Changes in relationships, familial roles and family dynamics
- Sexuality and intimacy
- Concerns about their family members and/or carers
- Decision-making [new 2016]

Offer the person information about sources of emotional and psychological support, including support groups and online forums. If needed, refer the person to counselling or psychology services for a specialist assessment and support. [new 2016]

During multidisciplinary team assessments and other appointments, discuss the psychological and emotional impact of MND with family members

and/or carers (as appropriate), and ask whether they have any psychological or social care support needs. Topics to discuss may include the following:

- Their understanding of MND and how it affects daily living
- Accepting and coping with the diagnosis and prognosis, including concerns and fears about the person with MND dying
- Adjusting to changes in their life
- Changes in relationships, familial roles and family dynamics, including their change to a carer role (if appropriate)
- Sexuality and intimacy
- Involvement in decision-making
- Impact on other family members and/or carers
- Their ability and willingness to provide personal care and operate equipment [new 2016]

Offer family members and/or carers (as appropriate) information about respite care and sources of emotional and psychological support, including support groups, online forums and counselling or psychology services. [new 2016]

A social care practitioner with knowledge of MND or rapidly progressive complex disabilities should discuss the person's needs and preferences for social care, and provide information and support for them to access the following:

- Personal care, ensuring there is continuity of care with familiar workers, so that wherever possible, personal care and support is carried out by workers known to the person and their family members and/or carers (as appropriate)
- Equipment and practical support (see "Equipment and Adaptations to Aid Activities of Daily Living and Mobility" below)
- Financial support and advice (for example, money management, how to access carers' and disability benefits and grants, continuing healthcare funding and funeral expenses)
- Support to engage in work, social activities and hobbies, such as access to social media and physical access to activities outside their home
- Respite care [new 2016]

Be aware that as MND progresses, people may develop communication problems and have difficulty accessing support or services. For example, they may be unable to access a call centre. Ensure people are given different ways of getting in touch with support or services, and a designated contact if possible. [new 2016]

### Planning for End of Life

Offer the person with MND the opportunity to discuss their preferences and concerns about care at the end of life at trigger points such as: at diagnosis, if there is a significant change in respiratory function, or if interventions such as gastrostomy or non-invasive ventilation are needed. Be sensitive about the timing of discussions and take into account the person's current communication ability, cognitive status and mental capacity. [new 2016]

Be prepared to discuss end of life issues whenever people wish to do so. [new 2016]

Provide support and advice on advance care planning for end of life. Topics to discuss may include:

- What could happen at the end of life, for example, how death may occur
- Providing anticipatory medicines in the home
- Advance care planning, including Advance Decisions to Refuse Treatment (ADRT) and Do Not Attempt Resuscitation (DNACPR) orders, and Lasting Power of Attorney
- How to ensure advance care plans will be available when needed, for example, including the information on the person's Summary Care Record
- When to involve specialist palliative care
- Areas that people might wish to plan for, such as:
  - What they want to happen (for example, their preferred place of death)
  - What they do not want to happen (for example, being admitted to hospital)
  - Who will represent their decisions, if necessary
  - What should happen if they develop an intercurrent illness [new 2016]

Think about discussing advance care planning with people at an earlier opportunity if you expect their communication ability, cognitive status or mental capacity to get worse. [new 2016]

Offer people the opportunity to talk about, and review any existing, ADRT, DNACPR orders and Lasting Power of Attorney when interventions

such as gastrostomy and non-invasive ventilation are planned. [new 2016]

Provide additional support as the end of life approaches, for example, additional social or nursing care to enable informal carers and family to reduce their carer responsibilities and spend time with the person with MND. [new 2016]

Towards the end of life, ensure there is prompt access to the following, if not already provided:

- A method of communication that meets the person's needs, such as an AAC system
- Specialist palliative care
- Equipment, if needed, such as syringe drivers, suction machines, riser–recliner chair, hospital bed, commode and hoist
- Anticipatory medicines, including opioids and benzodiazepines to treat breathlessness, and antimuscarinic medicines to treat problematic saliva and respiratory secretions [new 2016]

Offer bereavement support to family members and/or carers (as appropriate). [new 2016]

### Managing Symptoms

#### Pharmacological Treatments for Muscle Problems

Discuss the available treatment options for muscle problems. Take into account the person's needs and preferences, and whether they have any difficulties taking medicine (for example, if they have problems swallowing). [new 2016]

Consider quinine<sup>1</sup> as first-line treatment for muscle cramps in people with MND. If quinine is not effective, not tolerated or contraindicated, consider baclofen<sup>1</sup> instead as second-line treatment. If baclofen is not effective, not tolerated or contraindicated, consider tizanidine<sup>1</sup>, dantrolene<sup>1</sup> or gabapentin<sup>1</sup>. [new 2016]

Consider baclofen, tizanidine, dantrolene<sup>1</sup> or gabapentin<sup>1</sup> to treat muscle stiffness, spasticity or increased tone in people with MND. If these treatments are not effective, not tolerated or contraindicated, consider referral to a specialist service for the treatment of severe spasticity. [new 2016]

Review the treatments for muscle problems during multidisciplinary team assessments, ask about how the person is finding the treatment, whether it is working and whether they have any adverse side effects. [new 2016]

#### Exercise Programmes

Consider an exercise programme for people with MND to:

- Maintain joint range of movement
- Prevent contractures
- Reduce stiffness and discomfort
- Optimise function and quality of life [new 2016]

Choose a programme that is appropriate to the person's level of function and tailored to their needs, abilities and preferences. Take into account factors such as postural needs and fatigue. The programme might be a resistance programme, an active-assisted programme or a passive programme. [new 2016]

Check that family members and/or carers (as appropriate) are willing and able to help with exercise programmes. [new 2016]

Give advice to the person and their family members and/or carers (as appropriate) about safe manual handling. [new 2016]

If a person needs orthoses to help with muscle problems, they should be referred to orthotics services without delay, and the orthoses should be provided without delay. [new 2016]

#### Saliva Problems

If a person with MND has problems with saliva, assess the volume and viscosity of the saliva and the person's respiratory function, swallowing, diet, posture and oral care. [new 2016]

If a person with MND has problems with drooling of saliva (sialorrhoea), provide advice on swallowing, diet, posture, positioning, oral care and suctioning. [new 2016]

Consider a trial of antimuscarinic medicine<sup>1</sup> as the first-line treatment for sialorrhoea in people with MND. [new 2016]

Consider glycopyrrolate<sup>1</sup> as the first-line treatment for sialorrhoea in people with MND who have cognitive impairment, because it has fewer central nervous system side effects. [new 2016]

If first-line treatment for sialorrhoea is not effective, not tolerated or contraindicated, consider referral to a specialist service for Botulinum toxin A<sup>1</sup>. [new 2016]

If a person with MND has thick, tenacious saliva:

- Review all current medicines, especially any treatments for sialorrhoea
- Provide advice on swallowing, diet, posture, positioning, oral care, suctioning and hydration
- Consider treatment with humidification, nebulisers and carbocisteine [new 2016]

#### Equipment and Adaptations to Aid Activities of Daily Living and Mobility

Healthcare professionals and social care practitioners, which will include physiotherapists and occupational therapists, should assess and anticipate changes in the person's daily living needs, taking into account the following:

Activities of daily living, including personal care, dressing and bathing, housework, shopping, food preparation, eating and drinking, and ability to continue with current work and usual activities

Mobility and avoiding falls and problems from loss of dexterity

The home environment and the need for adaptations

The need for assistive technology, such as environmental control systems [new 2016]

Provide equipment and adaptations that meet the person's needs without delay, so that people can participate in activities of daily living and maintain their quality of life as much as possible. [new 2016]

Refer people to specialist services without delay if assistive technology such as environmental control systems is needed. People should be assessed and assistive technology provided without delay. [new 2016]

Refer people to wheelchair services without delay if needed. Wheelchair needs should be assessed and a manual and/or powered wheelchair that meets the person's needs should be provided without delay. [new 2016]

Ensure that equipment, adaptations, daily living aids, assistive technology and wheelchairs meet the changing needs of the person and their family and/or carers (as appropriate) to maximise mobility and participation in activities of daily living. [new 2016]

Ensure regular, ongoing monitoring of the person's mobility and daily life needs and abilities as MND progresses. Regularly review their ability to use equipment and to adapt equipment as necessary. [new 2016]

Healthcare professionals, social care practitioners and other services providing equipment should liaise to ensure that all equipment provided can be integrated, for example, integrating AAC aids and devices and environmental control systems with wheelchairs. [new 2016]

Enable prompt access and assessment for funding for home adaptation. If the person is not eligible for funding, continue to offer information and support in arranging home environment adaptations. [new 2016]

#### Nutrition and Gastrostomy

Please also refer to the recommendations in NICE's guideline on [nutrition support for adults](#) .

At diagnosis and at multidisciplinary team assessments, or if there are any concerns about weight, nutrition or swallowing, assess the person's weight, diet, nutritional intake, fluid intake, hydration, oral health, feeding, drinking and swallowing, and offer support, advice and interventions as needed. [new 2016]

Assess the person's diet, hydration, nutritional intake and fluid intake by taking into account:

- Fluids and food intake versus nutritional and hydration needs
- Nutritional supplements, if needed
- Appetite and thirst
- Gastrointestinal symptoms, such as nausea or constipation



- Causes of reduced oral intake (for example, swallowing difficulties, limb weakness or the possibility of low mood or depression causing loss of appetite) [new 2016]

Assess the person's ability to eat and drink by taking into account:

- The need for eating and drinking aids and altered utensils to help them take food from the plate to their mouth
- The need for help with food and drink preparation
- Advice and aids for positioning, seating and posture while eating and drinking
- Dealing with social situations (for example, eating out) [new 2016]

Arrange for a clinical swallowing assessment if swallowing problems are suspected. [new 2016]

Assess and manage factors that may contribute to problems with swallowing, such as:

- Positioning
- Seating
- The need to modify food and drink consistency and palatability
- Respiratory symptoms and risk of aspiration and/or choking
- Fear of choking and psychological considerations (for example, wanting to eat and drink without assistance in social situations) [new 2016]

Discuss gastrostomy at an early stage, and at regular intervals as MND progresses, taking into account the person's preferences and issues, such as ability to swallow, weight loss, respiratory function, effort of feeding and drinking and risk of choking. Be aware that some people will not want to have a gastrostomy. [new 2016]

Explain the benefits of early placement of a gastrostomy, and the possible risks of a late gastrostomy (for example, low critical body mass, respiratory complications, risk of dehydration, different methods of insertion, and a higher risk of mortality and procedural complications). [new 2016]

If a person is referred for a gastrostomy, it should take place without unnecessary delay. [new 2016]

Pay particular attention to the nutritional and hydration needs of people with MND who have frontotemporal dementia and who lack mental capacity. The multidisciplinary team assessment should include the support they need from carers, and their ability to understand the risks of swallowing difficulties. [new 2016]

Before a decision is made on the use of gastrostomy for a person with MND who has frontotemporal dementia, the neurologist from the multidisciplinary team should assess the following:

- The person's ability to make decisions and to give consent<sup>2</sup>
- The severity of frontotemporal dementia and cognitive problems
- Whether the person is likely to accept and cope with treatment

Discuss with the person's family members and/or carers (as appropriate; with the person's consent if they have the ability to give it). [new 2016]

## Communication

When assessing speech and communication needs during multidisciplinary team assessments and other appointments, discuss face-to-face and remote communication, for example, using the telephone, email, the Internet and social media. Ensure that the assessment and review is carried out by a speech and language therapist without delay. [new 2016]

Provide AAC equipment that meets the needs of the person without delay to maximise participation in activities of daily living and maintain quality of life. The use of both low-level technologies, for example, alphabet, word or picture boards and high-level technologies, for example, personal computer (PC) or tablet-based voice output communication aids may be helpful. Review the person's communication needs during multidisciplinary team assessments. [new 2016]

Liaise with, or refer the person with MND to, a specialised National Health Service (NHS) AAC hub if complex high technology AAC equipment (for example, eye gaze access) is needed or is likely to be needed. [new 2016]

Involve other healthcare professionals, such as occupational therapists, to ensure that AAC equipment is integrated with other assistive technologies, such as environmental control systems and personal computers or tablets. [new 2016]

Ensure regular, ongoing monitoring of the person's communication needs and abilities as MND progresses, and review their ability to use AAC equipment. Reassess and liaise with a specialised NHS AAC hub if needed. [new 2016]

Provide ongoing support and training for the person with MND, and their family members and/or carers (as appropriate), in using AAC equipment and other communication strategies. [new 2016]

### Respiratory Function and Respiratory Symptoms

Assess and monitor the person's respiratory function and symptoms. Treat people with MND and worsening respiratory impairment for reversible causes (for example, respiratory tract infections or secretion problems) before considering other treatments. [new 2016]

Offer non-invasive ventilation as treatment for people with respiratory impairment (see "Non-invasive Ventilation" below). Decisions to offer non-invasive ventilation should be made by the multidisciplinary team in conjunction with the respiratory ventilation service, and the person (see recommendations Under "Organisation of Care" above). [new 2016]

Consider urgent introduction of non-invasive ventilation for people with MND who develop worsening respiratory impairment and are not already using non-invasive ventilation. [new 2016]

Consider opioids<sup>1</sup> as an option to relieve symptoms of breathlessness. Take into account the route of administration and acquisition cost of medicines. [new 2016]

Consider benzodiazepines<sup>1</sup> to manage breathlessness that is exacerbated by anxiety. Take into account the route of administration and acquisition cost of medicines. [new 2016]

### Cough Effectiveness

Offer cough augmentation techniques such as manual assisted cough to people with MND who cannot cough effectively. [new 2016]

Consider unassisted breath stacking and/or manual assisted cough as the first-line treatment for people with MND who have an ineffective cough. [new 2016]

For people with bulbar dysfunction, or whose cough is ineffective with unassisted breath stacking, consider assisted breath stacking (for example, using a lung volume recruitment bag). [new 2016]

Consider a mechanical cough assist device if assisted breath stacking is not effective, and/or during a respiratory tract infection. [new 2016]

### Non-invasive Ventilation

#### Information and Support about Non-invasive Ventilation

Offer to discuss the possible use of non-invasive ventilation with the person and (if the person agrees) their family and carers, at an appropriate time and in a sensitive manner. This may be at one or more of the following times:

- Soon after MND is first diagnosed
- When monitoring respiratory function
- When respiratory function deteriorates
- If the person asks for information [2010]

Discussions about non-invasive ventilation should be appropriate to the stage of the person's illness, carried out in a sensitive manner and include information on:

- The possible symptoms and signs of respiratory impairment (see Box 1 below)
- The purpose, nature and timing of respiratory function tests, and explanations of the test results
- How non-invasive ventilation (as a treatment option) can improve symptoms associated with respiratory impairment and can be life prolonging, but does not stop progression of the underlying disease [2010, amended 2016]

When discussing non-invasive ventilation, explain the different ways that people can manage their breathlessness symptoms. This should include:

- Non-invasive ventilation, and its advantages and disadvantages
- Using non-invasive ventilation at different points in the course of the person's lifetime
- The possibility of the person becoming dependent on non-invasive ventilation

- Options for treating any infections
- Support and information on how to recognise and cope with a distressing situation
- The role of medication for breathing problems
- Psychological techniques and support [new 2016]

Check that the person thinking about non-invasive ventilation:

- Understands what non-invasive ventilation is and what it can achieve
- Recognises the need for regular review
- Has enough information about non-invasive ventilation and other options for breathing problems to make decisions about how and when to use it
- Understands possible problems with compatibility with other equipment, for example, eye gaze access systems [new 2016]

Explain that non-invasive ventilation can be stopped at any time. Reassure people that they can ask for help and advice if they need it, especially if they are dependent on non-invasive ventilation for 24 hours a day, or become distressed when attempting to stop it. Inform people that medicines can be used to alleviate symptoms (see recommendation above). [new 2016]

Ensure that families and carers:

- Have an initial assessment if the person they care for decides to use non-invasive ventilation, which should include:
  - Their ability and willingness to assist in providing non-invasive ventilation
  - Their training needs
- Have the opportunity to discuss any concerns they may have with members of the multidisciplinary team, the respiratory ventilation service and/or other healthcare professionals [2010]

## Identification and Assessment of Respiratory Impairment

### *Symptoms and Signs*

Monitor the symptoms and signs listed in box 1 to detect potential respiratory impairment. [2010, amended 2016]

#### Box 1. Symptoms and Signs of Potential Respiratory Impairment

Symptoms	Signs
Breathlessness	Increased respiratory rate
Orthopnoea	Shallow breathing
Recurrent chest infections	Weak cough <sup>1</sup>
Disturbed sleep	Weak sniff
Non-refreshing sleep	Abdominal paradox (inward movement of the abdomen during inspiration)
Nightmares	Use of accessory muscles of respiration
Daytime sleepiness	Reduced chest expansion on maximal inspiration
Poor concentration and/or memory	
Confusion	
Hallucinations	
Morning headaches	
Fatigue	
Poor appetite	
<sup>1</sup> Weak cough could be assessed by measuring peak cough flow	

## *Respiratory Function Tests*

As part of the initial assessment to diagnose MND, or soon after diagnosis, a healthcare professional from the multidisciplinary team who has appropriate competencies should perform the following tests (or arrange for them to be performed) to establish the person's baseline respiratory function:

- Oxygen saturation measured by pulse oximetry (SpO<sub>2</sub>):
  - This should be a single measurement of SpO<sub>2</sub> with the person at rest and breathing room air.
  - If it is not possible to perform pulse oximetry locally, refer the person to a respiratory ventilation service.

Then one or both of the following:

- Forced vital capacity (FVC) or vital capacity (VC)<sup>3</sup>
- Sniff nasal inspiratory pressure (SNIP) and/or maximal inspiratory pressure (MIP) [2010]

If the person has severe bulbar impairment or severe cognitive problems that may be related to respiratory impairment:

- Ensure that SpO<sub>2</sub> is measured (at rest and breathing room air).
- Do not perform the other respiratory function tests (FVC, VC, SNIP and MIP) if interfaces are not suitable for the person. [2010]

A healthcare professional with appropriate competencies should perform the respiratory function tests every 2 to 3 months, although tests may be performed more or less often depending on:

- Whether there are any symptoms and signs of respiratory impairment (see box 1 above)
- The rate of progression of MND
- The person's preference and circumstances [2010, amended 2016]

Perform arterial or capillary blood gas analysis if the person's SpO<sub>2</sub> (measured at rest and breathing room air):

- Is less than or equal to 92% if they have known lung disease
- Is less than or equal to 94% if they do not have lung disease

If it is not possible to perform arterial or capillary blood gas analysis locally, refer the person to a respiratory ventilation service. [2010]

If the person's SpO<sub>2</sub> (measured at rest and breathing room air) is greater than 94%, or 92% for those with lung disease, but they have sleep-related respiratory symptoms:

- Consider referring them to a respiratory ventilation service for continuous nocturnal (overnight) oximetry and/or a limited sleep study and
- Discuss both the impact of respiratory impairment and treatment options with the patient and (if the person agrees) their family and carers [2010]

If the person's arterial partial pressure of carbon dioxide (PaCO<sub>2</sub>) is greater than 6 kPa:

- Refer them urgently to a respiratory ventilation service (to be seen within 1 week) and
- Explain the reasons for and implications of the urgent referral to the person and (if the person agrees) their family and carers [2010]

If the person's PaCO<sub>2</sub> is less than or equal to 6 kPa but they have any symptoms or signs of respiratory impairment, particularly orthopnoea (see recommendation below):

- Refer them to a respiratory ventilation service for nocturnal (overnight) oximetry and/or a limited sleep study and
- Discuss both the impact of respiratory impairment and treatment options with the person and (if the person agrees) their family and/or carers (as appropriate) [2010]

If any of the results listed in Box 2 below is obtained, discuss with the person and (if appropriate) their family and carers:

- Their respiratory impairment
- Their treatment options
- Possible referral to a respiratory ventilation service for further assessment based on discussion with the person, and their wishes [2010, amended 2016]

## Box 2. Results of Respiratory Function Tests

Forced Vital Capacity (FVC) or Vital Capacity (VC)	Sniff Nasal Inspiratory Pressure (SNIP) and/or Maximal Inspiratory Pressure (MIP) (if both tests are performed, base the assessment on the better respiratory function reading)
FVC or VC less than 50% of predicted value	SNIP or MIP less than 40 cmH <sub>2</sub> O
FVC or VC less than 80% of predicted value plus any symptoms or signs of respiratory impairment (see recommendation above), particularly orthopnoea	SNIP or MIP less than 65 cmH <sub>2</sub> O for men or 55 cmH <sub>2</sub> O for women plus any symptoms or signs of respiratory impairment (see recommendation above), particularly orthopnoea  Repeated regular tests show a rate of decrease of SNIP or MIP of more than 10 cm H <sub>2</sub> O per 3 months

### *People with a Diagnosis of Frontotemporal Dementia*

Base decisions on respiratory function tests for a person with a diagnosis of frontotemporal dementia on considerations specific to their needs and circumstances, such as:

- Their ability to give consent<sup>2</sup>
- Their understanding of the tests
- Their tolerance of the tests and willingness to undertake them
- The impact on their family and carers
- Whether they are capable of receiving non-invasive ventilation [2010, amended 2016]

### Non-invasive Ventilation for Treatment of Respiratory Impairment in People with MND

Offer a trial of non-invasive ventilation if the person's symptoms and signs and the results of the respiratory function tests indicate that the person is likely to benefit from the treatment. [2010, amended 2016]

Consider a trial of non-invasive ventilation for a person who has severe bulbar impairment or severe cognitive problems that may be related to respiratory impairment only if they may benefit from an improvement in sleep-related symptoms or correction of hypoventilation. [2010, amended 2016]

Before starting non-invasive ventilation, the multidisciplinary team together with the respiratory ventilation service should carry out and coordinate a patient-centred risk assessment, after discussion with the person and their family and carers. This should consider:

- The most appropriate type of non-invasive ventilator and interfaces, based on the person's needs and lifestyle factors and safety
- The person's tolerance of the treatment
- The risk, and possible consequences, of ventilator failure
- The power supply required, including battery back-up
- How easily the person can get to hospital
- Risks associated with travelling away from home (especially abroad)
- Whether a humidifier is required
- Issues relating to secretion management
- The availability of carers [2010]

Before starting non-invasive ventilation, the multidisciplinary team together with the respiratory ventilation service should prepare a comprehensive care plan, after discussion with the person and their family and carers (who should be offered a copy of the plan). This should cover:

- Long-term support provided by the multidisciplinary team
- The initial frequency of respiratory function tests and monitoring of respiratory impairment
- The frequency of clinical reviews of symptomatic and physiological changes
- The provision of carers
- Arrangements for device maintenance and 24-hour emergency clinical and technical support

- Secretion management and respiratory physiotherapy assessment, including cough augmentation (if required)
- Training in and support for the use of non-invasive ventilation for the person and their family and carers
- Regular opportunities to discuss the person's wishes in relation to continuing or withdrawing non-invasive ventilation [2010, amended 2016]

When starting non-invasive ventilation:

- Perform initial acclimatisation during the day when the person is awake
- Usually start regular treatment at night, before and during sleep
- Gradually build up the person's hours of use as necessary [2010]

Continue non-invasive ventilation if the clinical reviews show:

- Symptomatic and/or physiological improvements for a person without severe bulbar impairment and without severe cognitive problems
- An improvement in sleep-related symptoms for a person with severe bulbar impairment or with severe cognitive problems that may be related to respiratory impairment [2010]

Provide the person and their family and/or carers (as appropriate) with support and assistance to manage non-invasive ventilation. This should include:

- Training on using non-invasive ventilation and ventilator interfaces, for example:
  - Emergency procedures
  - Night-time assistance if the person is unable to use the equipment independently (for example, emergency removal or replacement of interfaces)
  - How to use the equipment with a wheelchair or other mobility aids if required
  - What to do if the equipment fails
- Assistance with secretion management
- Information on general palliative strategies
- An offer of ongoing emotional and psychological support for the person and their family and carers [2010, amended 2016]

Discuss all decisions to continue or withdraw non-invasive ventilation with the person and (if the person agrees) their family and carers. [2010]

Before a decision is made on the use of non-invasive ventilation for a person with a diagnosis of frontotemporal dementia, the multidisciplinary team together with the respiratory ventilation service should carry out an assessment that includes:

- The person's capacity to make decisions and to give consent<sup>2</sup>
- The severity of dementia and cognitive problems
- Whether the person is likely to accept treatment
- Whether the person is likely to achieve improvements in sleep-related symptoms and/or behavioural improvements
- A discussion with the person's family and/or carers (with the person's consent if they have the capacity to give it) [2010, amended 2016]

Consider prescribing medicines to help ease breathlessness that people using non-invasive ventilation can take on an 'as-needed' basis at home, for example, opioids<sup>1</sup> or benzodiazepines<sup>1</sup>. [new 2016]

Inform services that may see the person in crisis situations, such as their GP and services that provide emergency or urgent care, that the person is using non-invasive ventilation. [new 2016]

### Stopping Non-invasive Ventilation

The healthcare professionals responsible for starting non-invasive ventilation treatment in people with MND should ensure that support is available for other healthcare professionals who may be involved if there is a plan to stop non-invasive ventilation, including the legal and ethical implications. [new 2016]

If a person on continuous non-invasive ventilation wishes to stop treatment, ensure that they have support from healthcare professionals with knowledge and expertise of:

- Stopping non-invasive ventilation
- The ventilator machine
- Palliative medicines (see the NGC summary of the NICE guideline [Care of dying adults in the last days of life](#))
- Supporting the person, family members and/or carers (as appropriate)

- Supporting other healthcare professionals involved with the person's care
- Legal and ethical frameworks and responsibilities [new 2016]

If a person on continuous non-invasive ventilation wishes to stop treatment, seek advice from healthcare professionals who have knowledge and experience of stopping non-invasive ventilation. [new 2016]

Healthcare professionals involved in stopping non-invasive ventilation should have up-to-date knowledge of the law regarding the Mental Capacity Act, DNACPR, ADRT orders, and Lasting Power of Attorney. [new 2016]

#### Footnotes

<sup>1</sup>At the time of publication (February 2016), these medicines did not have a UK marketing authorisation for this indication. The prescriber should follow relevant professional guidance, taking full responsibility for the decision. Informed consent should be obtained and documented. See the General Medical Council's [Prescribing guidance: prescribing unlicensed medicines](#)  for further information.

<sup>2</sup>See [Mental Capacity Act 2005](#) .

<sup>3</sup>The difference between the measurement of vital capacity and forced vital capacity is very subtle and so either can be used.

#### Definitions

##### Strength of Recommendations

Some recommendations can be made with more certainty than others. The Guideline Committee makes a recommendation based on the trade-off between the benefits and harms of an intervention, taking into account the quality of the underpinning evidence. For some interventions, the Guideline Committee is confident that, given the information it has looked at, most patients would choose the intervention. The wording used in the recommendations in this guideline denotes the certainty with which the recommendation is made (the strength of the recommendation).

##### *Interventions That Must (or Must Not) Be Used*

The Guideline Committee usually uses 'must' or 'must not' only if there is a legal duty to apply the recommendation. Occasionally the Guideline Committee uses 'must' (or 'must not') if the consequences of not following the recommendation could be extremely serious or potentially life threatening.

##### *Interventions That Should (or Should Not) Be Used – a 'Strong' Recommendation*

The Guideline Committee uses 'offer' (and similar words such as 'refer' or 'advise') when confident that, for the vast majority of patients, an intervention will do more good than harm, and be cost effective. The Guideline Committee uses similar forms of words (for example, 'Do not offer...') when confident that an intervention will not be of benefit for most patients.

##### *Interventions That Could Be Used*

The Guideline Committee uses 'consider' when confident that an intervention will do more good than harm for most patients, and be cost effective, but other options may be similarly cost effective. The choice of intervention, and whether or not to have the intervention at all, is more likely to depend on the patient's values and preferences than for a strong recommendation, and so the healthcare professional should spend more time considering and discussing the options with the patient.

## Clinical Algorithm(s)

A National Institute for Health and Care Excellence (NICE) pathway titled "Motor neurone disease overview" is provided on the [NICE Web site](#) .

## Scope

## Disease/Condition(s)

- Motor neurone disease (MND) (amyotrophic lateral sclerosis [ALS])

- Frontotemporal dementia

## Guideline Category

Counseling

Diagnosis

Evaluation

Management

Treatment

## Clinical Specialty

Family Practice

Neurology

Nutrition

Physical Medicine and Rehabilitation

Speech-Language Pathology

## Intended Users

Advanced Practice Nurses

Dietitians

Health Care Providers

Health Plans

Hospitals

Managed Care Organizations

Nurses

Occupational Therapists

Patients

Physical Therapists

Physician Assistants

Physicians

Psychologists/Non-physician Behavioral Health Clinicians

Public Health Departments

Respiratory Care Practitioners

Social Workers

Speech-Language Pathologists



## Guideline Objective(s)

- To consider the clinical- and cost-effectiveness evidence for the care of people with motor neurone disease (MND) from the time of diagnosis, including communication of the diagnosis
- To provide recommendations on monitoring of disease progression, management of symptoms (in particular muscle weakness, excess secretions, breathing and nutrition problems), ongoing support and services, mobility, emotional and psychological changes, and preparation for end of life
- To determine the best way to organise the care and management of people with MND

## Target Population

Adults (aged 18 and over) with motor neurone disease (MND)

Note: People with frontotemporal dementia are considered as a separate patient subgroup. Groups that will not be covered are: children and young people (under 18 years), adults with other neurodegenerative disorders who do not have MND, and people with Kennedy's disease.

## Interventions and Practices Considered

1. Recognition of symptoms of motor neurone disease (MND)
2. Referral to neurologist
3. Providing information and support to patients with MND and their families and carers
4. Cognitive assessments
5. Taking into account prognostic factors
6. Organisation of care around an MND multidisciplinary team
7. Providing psychological and social care support
8. Planning for end of life
9. Managing symptoms
  - Pharmacological treatments for muscle problems (e.g., quinine, baclofen, tizanidine, dantrolene, gabapentin)
  - Exercise programs
  - Assessing and managing saliva problems (e.g., antimuscarinic agent such as glycopyrrolate for sialorrhoea)
10. Equipment and adaptations to aid activities of daily living and mobility
11. Assessing the person's diet, hydration, nutritional intake, fluid intake, and ability to eat and drink
12. Referral for gastrostomy
13. Assessing and addressing communication needs such as provision of alternative and augmentative communication (AAC) services
14. Assessing and monitoring the person's respiratory function and symptoms
15. Opioids and benzodiazepines for breathlessness
16. Cough augmentation techniques
17. Providing non-invasive ventilation and support in its use
18. Identification and assessment of respiratory impairment (respiratory function tests)
19. Stopping non-invasive ventilation

## Major Outcomes Considered

- Health-related quality of life
- Patient and carer-reported outcomes, for example, symptoms, satisfaction, and pain
- Hospital admissions (including unplanned admissions)
- Mobility
- Survival/time to death
- Reduction of muscle weakness, increased tone, and muscle cramps
- Aspiration pneumonia
- Change in nutritional status
- Mortality related to procedures
- Cost-effectiveness

# Methodology

## Methods Used to Collect/Select the Evidence

Hand-searches of Published Literature (Primary Sources)

Hand-searches of Published Literature (Secondary Sources)

Searches of Electronic Databases

## Description of Methods Used to Collect/Select the Evidence

Note from the National Guideline Clearinghouse (NGC): This guideline was developed by the National Clinical Guideline Centre (NCGC) on behalf of the National Institute for Health and Care Excellence (NICE). See the "Availability of Companion Documents" field for the full version of this guidance and related appendices.

### Developing the Review Questions and Outcomes

Review questions were developed using a PICO framework (patient, intervention, comparison and outcome) for intervention reviews; using a framework of population, index tests, reference standard and target condition for reviews of diagnostic test accuracy; and using population, presence or absence of factors under investigation (for example prognostic factors) and outcomes for prognostic reviews.

This use of a framework guided the literature searching process, critical appraisal and synthesis of evidence, and facilitated the development of recommendations by the Guideline Development Group (GDG). The review questions were drafted by the NCGC technical team and refined and validated by the GDG. The questions were based on the key clinical areas identified in the scope (see Appendix A) were identified.

Full literature searches, critical appraisals and evidence reviews were completed for all the specified review questions.

### Searching for Evidence

#### Clinical Literature Search

The aim of the literature search was to systematically identify all published clinical evidence relevant to the review questions. Searches were undertaken according to the parameters stipulated within the NICE guidelines manual (see the "Availability of Companion Documents" field). Databases were searched using relevant medical subject headings, free-text terms and study-type filters where appropriate. Foreign language studies were not reviewed and, where possible, searches were restricted to articles published in the English language. All searches were conducted in MEDLINE, EMBASE, and The Cochrane Library. Additional subject specific databases (CINAHL and PsycINFO) were used for some questions. All searches were updated on 18 May 2015. No papers published after this date were considered except for one paper. The GDG was aware that this paper was due for publication soon after the cut-off date and wished to include it in the cough augmentation question.

Search strategies were quality assured by cross-checking reference lists of highly relevant papers, analysing search strategies in other systematic reviews, and asking GDG members to highlight any additional studies. The questions, the study types applied, the databases searched and the years covered can be found in Appendix F.

The titles and abstracts of records retrieved by the searches were sifted for relevance, with potentially significant publications obtained in full text. These were assessed against the inclusion criteria.

During the scoping stage, a search was conducted for guidelines and reports on the Web sites listed below from organisations relevant to the topic. Searching for unpublished literature was not undertaken. All references sent by stakeholders were considered.

- Guidelines International Network database ([www.g-i-n.net](http://www.g-i-n.net) )
- NGC ([www.guideline.gov](http://www.guideline.gov) )
- NICE ([www.nice.org.uk](http://www.nice.org.uk) )
- National Institutes of Health Consensus Development Program ([consensus.nih.gov](http://consensus.nih.gov) )
- National Health Service (NHS) Evidence Search ([www.evidence.nhs.uk](http://www.evidence.nhs.uk) )

#### Health Economic Literature Search

Systematic literature searches were also undertaken to identify health economic evidence within published literature relevant to the review questions. The evidence was identified by conducting a broad search relating to motor neurone disease (MND) in the NHS Economic Evaluation Database (NHS EED), the Health Technology Assessment database (HTA) and the Health Economic Evaluations Database (HEED) with no date restrictions. Additionally, the search was run on MEDLINE and EMBASE using a specific economic filter, from 2012, to ensure recent publications that had not yet been indexed by the economic databases were identified. Foreign language studies were not reviewed and, where possible, searches were restricted to articles published in the English language.

The health economic search strategies are included in Appendix F. All searches were updated on 18 May 2015. No papers published after this date were considered.

### Evidence Gathering and Analysis

The tasks of the research fellow are listed below and described in further detail in Sections 4.3.1 to 4.3.6. The research fellow:

- Identified potentially relevant studies for each review question from the relevant search results by reviewing titles and abstracts, and deciding which should be ordered as full papers. Full papers were then obtained.
- Reviewed full papers against pre-specified inclusion/exclusion criteria to identify studies that addressed the review question in the appropriate population, and reported on outcomes of interest (see Appendix C).

### Inclusion and Exclusion Criteria

The inclusion and exclusion of studies was based on the criteria defined in the review protocols (see Appendix C). Excluded studies by review question (with the reasons for their exclusion) are listed in Appendix K. The GDG was consulted about any uncertainty regarding inclusion or exclusion.

The key population inclusion criterion was:

- Adults (aged 18 and over) with motor neurone disease

The key population exclusion criteria were:

- Children and young people (under 18 years)
- Adults with other neurodegenerative disorders who do not have MND
- People diagnosed with Kennedy's disease

Conference abstracts were not automatically excluded from any review. The abstracts were initially assessed against the inclusion criteria for the review question and further processed where a full publication was not available. If the abstracts were included, the authors were contacted for further information. No relevant conference abstracts were identified for this guideline. Literature reviews, posters, letters, editorials, comment articles, unpublished studies and studies not in English were excluded.

### Type of Studies

Randomised trials, non-randomised trials, and observational studies (including diagnostic and prognostic studies) were included in the evidence reviews as appropriate. Qualitative reviews were included where relevant to a particular question, and specified in the protocol.

For most intervention reviews in this guideline, parallel randomised controlled trials (RCTs) were included because they are considered the most robust type of study design that could produce an unbiased estimate of the intervention effects. Crossover RCTs were appropriate for the questions 'What is the clinical- and cost-effectiveness of interventions for saliva management in people with MND?' and 'What is the clinical and cost-effectiveness of cough augmentation techniques for people with MND who have an ineffective cough?' If non-randomised studies were appropriate for inclusion, that is, non-drug trials with no randomised evidence, the GDG identified a priori in the protocol that the variables must either be equivalent at baseline or that the analysis had to adjust for any baseline differences. If the study did not fulfil either criterion it was excluded. Please refer to Appendix C for full details of the study design of studies selected for each review question.

For prognostic reviews, prospective and retrospective cohort and case-control studies were included.

### Evidence of Cost-effectiveness

The GDG is required to make decisions based on the best available evidence of both clinical and cost-effectiveness. Guideline recommendations should be based on the expected costs of the different options in relation to their expected health benefits (that is, their 'cost-effectiveness') rather than the total implementation cost. Thus, if the evidence suggests that a strategy provides significant health benefits at an acceptable cost per patient

treated, it should be recommended even if it would be expensive to implement across the whole population.

Evidence on cost-effectiveness related to the key clinical issues being addressed in the guideline was sought. The health economist:

- Undertook a systematic review of the published economic literature
- Undertook new cost-effectiveness analysis in priority areas

Literature Review

The health economist:

- Identified potentially relevant studies for each review question from the economic search results by reviewing titles and abstracts. Full papers were then obtained.
- Reviewed full papers against pre-specified inclusion/exclusion criteria to identify relevant studies (see below for details)

*Inclusion and Exclusion Criteria*

Full economic evaluations (studies comparing costs and health consequences of alternative courses of action: cost–utility, cost-effectiveness, cost–benefit and cost–consequence analyses) and comparative costing studies that addressed the review question in the relevant population were considered potentially applicable as economic evidence.

Studies that only reported cost per hospital (not per patient) or only reported average cost effectiveness without disaggregated costs and effects were excluded. Literature reviews, abstracts, posters, letters, editorials, comment articles, unpublished studies and studies not in English were excluded. Studies published before 1999 and studies from non-Organisation for Economic Co-operation and Development (OECD) countries or the USA were also excluded.

Remaining studies were prioritised for inclusion based on their relative applicability to the development of this guideline and the study limitations. For example, if a high quality, directly applicable UK analysis was available, then other less relevant studies may not have been included. Where exclusions occurred on this basis, this is noted in the relevant section.

For more details about the assessment of applicability and methodological quality, see Table 7 in the full version of the guideline and the economic evaluation checklist (Appendix G of the NICE guidelines manual 2012) and the health economics review protocol in Appendix C of the full guideline appendices.

When no relevant economic analysis was found from the economic literature review, relevant UK NHS unit costs related to the compared interventions were presented to the GDG to inform the possible economic implications of the recommendations being made.

Number of Source Documents

See Appendix D: Clinical Article Selection and Appendix E: Economic Article Selection (see the "Availability of Companion Documents" field) for detailed flow charts on the article selection process, including total number of records identified through database searching, records screened, records excluded, full-text articles assessed for eligibility, studies included in review, and studies excluded from review.

Methods Used to Assess the Quality and Strength of the Evidence

Weighting According to a Rating Scheme (Scheme Given)

Rating Scheme for the Strength of the Evidence

Overall Quality of Outcome Evidence in Grading of Recommendations Assessment, Development and Evaluation (GRADE)

Level	Description
High	Further research is very unlikely to change confidence in the estimate of effect.
Moderate	Further research is likely to have an important impact on confidence in the estimate of effect and may change the estimate.
Low	Further research is very likely to have an important impact on confidence in the estimate of effect and is likely to change the

Level	estimate Description
Very Low	Any estimate of effect is very uncertain.

## Methods Used to Analyze the Evidence

Meta-Analysis

Review of Published Meta-Analyses

Systematic Review with Evidence Tables

## Description of the Methods Used to Analyze the Evidence

Note from the National Guideline Clearinghouse (NGC): This guideline was developed by the National Clinical Guideline Centre (NCGC) on behalf of the National Institute for Health and Care Excellence (NICE). See the "Availability of Companion Documents" field for the full version of this guidance and related appendices.

### Evidence Gathering and Analysis

The tasks of the research fellow are listed below and described in further detail in the full version of the guideline. The research fellow:

- Critically appraised relevant studies using the appropriate study design checklists as specified in The Guidelines Manual (NICE [2012]) (see the "Availability of Companion Documents" field).
- Critically appraised relevant studies with a prognostic or qualitative study design using the NCGC checklist.
- Extracted key information about interventional study methods and results using Evibase, NCGC purpose-built software. Evibase produces summary evidence tables, with critical appraisal ratings. Key information about non-interventional study methods and results were manually extracted onto standard evidence tables and critically appraised separately (see Appendix G for evidence tables).
- Generated summaries of the evidence by outcome. Outcome data were combined, analysed and reported according to study design:
  - i. Randomised data were meta analysed where appropriate and reported in Grading of Recommendations Assessment, Development and Evaluation (GRADE) profiles.
  - ii. Observational data were presented separately in GRADE profiles.
  - iii. Prognostic data were meta-analysed where appropriate and reported in GRADE profiles.
  - iv. Qualitative data were summarised across studies where appropriate and reported in themes.
- A sample of a minimum of 10% of the abstract lists of each review was conducted. All of the evidence reviews were quality assured by a senior research fellow. This included checking:
  - i. Papers were included or excluded appropriately
  - ii. A sample of the data extractions
  - iii. Correct methods were used to synthesise data
  - iv. A sample of the risk of bias assessment

### Methods of Combining Evidence

#### *Data Synthesis for Intervention Reviews*

Where possible, meta-analyses were conducted to combine the data from the studies for each of the outcomes in the review question using RevMan5 software.

Analyses were stratified for by relevant populations such as 'people with cognitive impairment including frontotemporal dementia', which meant that different studies with predominant cognitive impairment strata were not combined and analysed together with studies that did not predominantly include this population. Stratification tended to vary by question, and this is documented in the individual question protocols (see Appendix C). If additional strata were used this led to sub-strata (for example, 2 stratification criteria would lead to 4 sub-strata categories, or 3 stratification criteria would lead to 9 sub-strata categories) which would be analysed separately.

See Section 4.3.3.1 of the full version of the guideline for details regarding analysis of different types of data including dichotomous outcomes, continuous outcomes, generic inverse variance, heterogeneity, and complex analysis/further analysis.

### *Data Synthesis for Prognostic Factor Reviews*

Odds ratios (ORs), risk ratios (RRs) or hazard ratios (HRs), with their 95% confidence intervals (95% CIs) for the effect of the pre-specified prognostic factors were extracted from the studies. Studies were only included if the risk factors pre-specified by the GDG were adjusted for each other using multivariate analysis.

### *Data Synthesis for Risk Prediction Rules*


Evidence reviews on risk prediction rules/tools results were presented separately for discrimination and calibration. The discrimination data was analysed according to the principles outlined under the section on data synthesis for diagnostic accuracy studies. Calibration data, for example  $R^2$ , if reported, was presented separately to the discrimination data. The results were presented for each study separately along with the quality rating for the study. Inconsistency and imprecision were not assessed.

### *Data Synthesis for Qualitative Reviews*

For each included paper, sub-themes were identified and linked to a generic theme. An example of a sub-theme identified by patients and carers is 'Subsequent feelings after diagnosis making sense of it' and this is linked to a broader generic theme of 'Coping with the diagnosis.' A summary evidence table of generic themes and underpinning sub-themes was then produced alongside the quality of the evidence. The methodological quality of each study was assessed by one reviewer using NCGC-modified NICE checklists and the quality of the evidence was assessed by a modified GRADE approach for each outcome. This took into account the applicability and theme saturation/sufficiency of the evidence. The evidence was graded 'applicable' if the evidence was directly applicable to the question, and graded partially applicable if it was related but not sufficiently. The theme was 'saturated' if the findings for a theme were based on a broad range of views, including quotes and experience from a range of people, and authors followed up enough people to have sufficient saturation of data. This was detailed in the accompanying footnotes. Grading of the evidence started at high and was downgraded by one increment if assessed as not applicable and downgraded one increment if the theme was not saturated.

### *Appraising the Quality of Evidence by Outcomes*

#### *Interventional Studies*

The evidence for outcomes from the included RCT and observational studies were evaluated and presented using an adaptation of the 'Grading of Recommendations Assessment, Development and Evaluation (GRADE) toolbox' developed by the international [GRADE working group](#) . The software (GRADEpro) developed by the GRADE working group was used to assess the quality of each outcome, taking into account individual study quality and the meta-analysis results.

Each outcome was first examined for each of the quality elements listed and defined in Table 2 in the full version of the guideline.

Details of how the 4 main quality elements (risk of bias, indirectness, inconsistency and imprecision) were appraised for each outcome are given in Section 4.3.4 of the full version of the guideline. Publication or other bias was only taken into consideration in the quality assessment if it was apparent.

#### Overall Grading of the Quality of Clinical Evidence

Once an outcome had been appraised for the main quality elements, an overall quality grade was calculated for that outcome. The scores from each of the main quality elements (0, -1 or -2) were summed to give a score that could be anything from 0 (the best possible) to -8 (the worst possible). However, scores were capped at -3. This final score was then applied to the starting grade that had originally been applied to the outcome by default, based on study design. For example, all RCTs started as HIGH and the overall quality became MODERATE, LOW or VERY LOW if the overall score was -1, -2 or -3 points respectively. The significance of these overall ratings is explained in the "Rating Scheme for the Strength of the Evidence" field. The reasons or criteria used for downgrading were specified in the footnotes of the GRADE tables.

On the other hand, observational interventional studies started at LOW, and so a score of -1 would be enough to take the grade to the lowest level of VERY LOW. Observational studies could, however, be upgraded if there was: a large magnitude of effect, a dose-response gradient, and if all plausible confounding would reduce a demonstrated effect.

See Tables 5 to 7 in the full version of the guideline for a description of quality elements for different types of studies.

#### *Assessing Clinical Importance*

The GDG assessed the evidence by outcome in order to determine if there was, or potentially was, a clinically important benefit, a clinically important harm or no clinically important difference between interventions. To facilitate this, binary outcomes were converted into absolute risk

differences (ARDs) using GRADEpro software: the median control group risk across studies was used to calculate the ARD and its 95% CI from the pooled risk ratio.

The assessment of clinical benefit, harm, or no benefit or harm was based on the point estimate of absolute effect for intervention studies which was standardised across the reviews. The GDG considered for most of the outcomes in the intervention reviews that if at least 100 participants per 1000 (10%) achieved (if positive) the outcome of interest in the intervention group compared to the comparison group then this intervention would be considered beneficial. The same point estimate but in the opposite direction would apply if the outcome was negative. However, the control group rate was always taken into consideration and smaller control group rates could identify a clinical benefit/harm for the intervention group at lower than 100 participants.

This assessment was carried out by the GDG for each critical outcome, and an evidence summary table was produced to compile the GDG's assessments of clinical importance per outcome, alongside the evidence quality and the uncertainty in the effect estimate (imprecision).

### Clinical Evidence Statements

Clinical evidence statements are summary statements that are presented after the GRADE profiles, summarising the key features of the clinical effectiveness evidence presented. The wording of the evidence statements reflects the certainty/uncertainty in the estimate of effect. The evidence statements were presented by outcome and encompassed the following key features of the evidence:

- The number of studies and the number of participants for a particular outcome
- An indication of the direction of clinical importance (if one treatment is beneficial or harmful compared to the other or whether there is no difference between the two tested treatments)
- A description of the overall quality of evidence (GRADE overall quality)

### Evidence of Cost-effectiveness

The GDG is required to make decisions based on the best available evidence of both clinical- and cost-effectiveness. Guideline recommendations should be based on the expected costs of the different options in relation to their expected health benefits (that is, their 'cost-effectiveness') rather than the total implementation cost. Thus, if the evidence suggests that a strategy provides significant health benefits at an acceptable cost per patient treated, it should be recommended even if it would be expensive to implement across the whole population.

Evidence on cost-effectiveness related to the key clinical issues being addressed in the guideline was sought. The health economist:

- Undertook a systematic review of the economic literature
- Undertook new cost-effectiveness analysis in priority areas

### Literature Review

The health economist:

- Identified potentially relevant studies for each review question from the economic search results by reviewing titles and abstracts. Full papers were then obtained.
- Reviewed full papers against prespecified inclusion and exclusion criteria to identify relevant studies (see below for details).
- Critically appraised relevant studies using the economic evaluations checklist as specified in the NICE guidelines manual.
- Extracted key information about the studies' methods and results into evidence tables (included in Appendix H).
- Generated summaries of the evidence in NICE economic evidence profiles (included in the relevant chapter for each review question) – see below for details.

### NICE Economic Evidence Profiles

The NICE economic evidence profile has been used to summarise cost and cost-effectiveness estimates. The economic evidence profile shows an assessment of applicability and methodological quality for each economic evaluation, with footnotes indicating the reasons for the assessment. These assessments were made by the health economist using the economic evaluation checklist from the NICE guidelines manual. It also shows the incremental costs, incremental effects (for example, quality-adjusted life years [QALYs]) and incremental cost-effectiveness ratio for the base case analysis in the evaluation, as well as information about the assessment of uncertainty in the analysis. See Table 7 in the full version of the guideline for more details.

If a non-UK study was included in the profile, the results were converted into pounds sterling using the appropriate purchasing power parity.

### Undertaking New Health Economic Analysis

As well as reviewing the published economic literature for each review question, as described above, new economic analysis was undertaken by the health economist in selected areas. Priority areas for new health economic analysis were agreed by the GDG after formation of the review questions and consideration of the available health economic evidence.

The GDG identified coordination of care as the highest priority area for original economic modelling. This question was chosen as it will impact every individual with motor neurone disease (MND) regardless of the type of MND they have or severity of symptoms, meaning it could have large resource implications. Secondly, it was expected that good evidence would exist that would allow a robust analysis to be undertaken.

The following general principles were adhered to in developing the cost-effectiveness analysis:

- Methods were consistent with the NICE reference case.
- The GDG was involved in the design of the model, selection of inputs and interpretation of the results.
- Model inputs were based on the systematic review of the clinical literature supplemented with other published data sources where possible.
- Where published data were not available, GDG expert opinion was used to populate the model.
- Model inputs and assumptions were reported fully and transparently.
- The results were subject to sensitivity analysis and limitations were discussed.
- The model was peer-reviewed by another health economist at the NCGC.

Full methods for the cost-effectiveness analysis for coordination of care are described in Appendix M.

#### Cost-effectiveness Criteria

NICE's report 'Social value judgements: principles for the development of NICE guidance' sets out the principles that GDGs should consider when judging whether an intervention offers good value for money. In general, an intervention was considered to be cost-effective if either of the following criteria applied (given that the estimate was considered plausible):

- The intervention dominated other relevant strategies (that is, it was both less costly in terms of resource use and more clinically effective compared with all the other relevant alternative strategies), or
- The intervention cost less than £20,000 per QALY gained compared with the next best strategy

If the GDG recommended an intervention that was estimated to cost more than £20,000 per QALY gained, or did not recommend one that was estimated to cost less than £20,000 per QALY gained, the reasons for this decision are discussed explicitly in the 'Recommendations and link to evidence' section of the relevant chapter, with reference to issues regarding the plausibility of the estimate or to the factors set out in 'Social value judgements: principles for the development of NICE guidance'.

If a study reported the cost per life year gained but not QALYs, the cost per QALY gained was estimated by multiplying by an appropriate utility estimate to aid interpretation. The estimated cost per QALY gained is reported in the economic evidence profile with a footnote detailing the life-years gained and the utility value used. When QALYs or life years gained are not used in the analysis, results are difficult to interpret unless one strategy dominates the others with respect to every relevant health outcome and cost.

#### In the Absence of Economic Evidence

When no relevant published studies were found, and a new analysis was not prioritised, the GDG made a qualitative judgement about cost-effectiveness by considering expected differences in resource use between options and relevant UK National Health Service (NHS) unit costs, alongside the results of the clinical review of effectiveness evidence.

The UK NHS costs reported in the guideline are those that were presented to the GDG and were correct at the time recommendations were drafted. They may have changed subsequently before the time of publication. However, we have no reason to believe they have changed substantially.

## Methods Used to Formulate the Recommendations

### Expert Consensus

## Description of Methods Used to Formulate the Recommendations

Note from the National Guideline Clearinghouse (NGC): This guideline was developed by the National Clinical Guideline Centre (NCGC) on



behalf of the National Institute for Health and Care Excellence (NICE). See the "Availability of Companion Documents" field for the full version of this guidance.

### Who Developed This Guideline?

A multidisciplinary Guideline Development Group (GDG) comprising health professionals and researchers as well as lay members developed this guideline.

The GDG was convened by the NCGC in accordance with guidance from NICE. The group met every 5 to 6 weeks during the development of the guideline.

Staff from the NCGC provided methodological support and guidance for the development process. The team working on the guideline included a project manager, systematic reviewers, health economists and information scientists. They undertook systematic searches of the literature, appraised the evidence, conducted meta-analysis and cost-effectiveness analysis where appropriate and drafted the guideline in collaboration with the GDG.

### Developing Recommendations

Over the course of the guideline development process, the GDG was presented with:

- Evidence tables of the clinical and economic evidence reviewed from the literature. All evidence tables are in Appendices G and H.
- Summaries of clinical and economic evidence and quality (as presented in Chapters 5 to 21 in the full version of the guideline)
- Forest plots (see Appendix J)
- A description of the methods and results of the cost-effectiveness analysis undertaken for the guideline (see Appendix M)

Recommendations were drafted on the basis of the GDG's interpretation of the available evidence, taking into account the balance of benefits, harms and costs between different courses of action. This was either done formally in an economic model, or informally. Firstly, the net benefit over harm (clinical effectiveness) was considered, focusing on the critical outcomes. When this was done informally, the GDG took into account the clinical benefits and harms when one intervention was compared with another. The assessment of net benefit was moderated by the importance placed on the outcomes (the GDG's values and preferences), and the confidence the GDG had in the evidence (evidence quality). Secondly, whether the net benefit justified any differences in costs was assessed.

When clinical and economic evidence was of poor quality, conflicting or absent, the GDG drafted recommendations based on their expert opinion. The considerations for making consensus-based recommendations include the balance between potential harms and benefits, the economic costs compared to the economic benefits, current practices, recommendations made in other relevant guidelines, patient preferences and equality issues. The consensus recommendations were agreed through discussions in the GDG. The GDG also considered whether the uncertainty was sufficient to justify delaying making a recommendation to await further research, taking into account the potential harm of failing to make a clear recommendation.

The GDG considered the 'strength' of recommendations. This takes into account the quality of the evidence but is conceptually different. Some recommendations are 'strong' in that the GDG believes that the vast majority of healthcare and other professionals and patients would choose a particular intervention if they considered the evidence in the same way that the GDG has. This is generally the case if the benefits clearly outweigh the harms for most people and the intervention is likely to be cost-effective. However, there is often a closer balance between benefits and harms, and some patients would not choose an intervention whereas others would. This may happen, for example, if some patients are particularly averse to some side effect and others are not. In these circumstances the recommendation is generally weaker, although it may be possible to make stronger recommendations about specific groups of patients.

The GDG focused on the following factors in agreeing the wording of the recommendations:

- The actions health professionals need to take
- The information readers need to know
- The strength of the recommendation (for example the word 'offer' was used for strong recommendations and 'consider' for weak recommendations)
- The involvement of patients (and their carers if needed) in decisions on treatment and care
- Consistency with NICE's standard advice on recommendations about drugs, waiting times and ineffective interventions

The main considerations specific to each recommendation are outlined in the 'Recommendations and link to evidence' sections within each chapter in the full version of the guideline.

# Rating Scheme for the Strength of the Recommendations

## Strength of Recommendations

Some recommendations can be made with more certainty than others. The Guideline Committee makes a recommendation based on the trade-off between the benefits and harms of an intervention, taking into account the quality of the underpinning evidence. For some interventions, the Guideline Committee is confident that, given the information it has looked at, most patients would choose the intervention. The wording used in the recommendations in this guideline denotes the certainty with which the recommendation is made (the strength of the recommendation).

### Interventions That Must (or Must Not) Be Used

The Guideline Committee usually uses 'must' or 'must not' only if there is a legal duty to apply the recommendation. Occasionally the Guideline Committee uses 'must' (or 'must not') if the consequences of not following the recommendation could be extremely serious or potentially life threatening.

### Interventions That Should (or Should Not) Be Used – a 'Strong' Recommendation

The Guideline Committee uses 'offer' (and similar words such as 'refer' or 'advise') when confident that, for the vast majority of patients, an intervention will do more good than harm, and be cost effective. The Guideline Committee uses similar forms of words (for example, 'Do not offer...') when confident that an intervention will not be of benefit for most patients.

### Interventions That Could Be Used

The Guideline Committee uses 'consider' when confident that an intervention will do more good than harm for most patients, and be cost effective, but other options may be similarly cost effective. The choice of intervention, and whether or not to have the intervention at all, is more likely to depend on the patient's values and preferences than for a strong recommendation, and so the healthcare professional should spend more time considering and discussing the options with the patient.

## Cost Analysis

Economic evidence is provided for each review question in the full version of the guideline (see the "Availability of Companion Documents" field).

See also the "Availability of Companion Documents" field for Appendix M: Cost-effectiveness Analysis: Multi-Disciplinary Care (see model overview and conclusions below).

## Model Overview/Methods

### Comparators

Only 2 comparators were considered in this economic evaluation:

- General care – currently when an individual is diagnosed with motor neurone disease (MND) the majority will continue to be reviewed in a general neurology clinic. The neurologist running this clinic would usually have a primary interest that is not MND. The individual would likely be reviewed once or twice a year where monitoring and discussion of future interventions would be discussed.
- Multi-disciplinary team (MDT) care – another type of care that some individuals with MND receive at diagnosis is delivered by a specialist MDT clinic. These clinics comprise of an extended team of specialists whose primary interest is MND. The individual will be regularly reviewed and monitored by this team.

Although the composition of specialists in an MDT could vary, there was no clinical evidence that specifically evaluated the increased effectiveness of each additional specialist in an MDT. Therefore the MDT composition in the model was the same as that used in the clinical studies.

### Approach to Modelling

The cost-effectiveness of MDT care was evaluated with the use of a discrete event simulation (DES) model. DESs treat time as a continuous variable and track costs and health outcomes over the course of a simulation. Within this simulation the individual will be exposed to a series of events that can occur at any timepoint throughout the simulation. These events will influence costs and health outcomes and might be re-occurring or only happen once (for example, death). The simulation ends once the individual has died or the model has reached its set time horizon. Time-to-event is the key parameter in DESs and these values are often characterised using exponential or Weibull distributions. This is further elaborated in section M.2.3 of Appendix M.

## Summary of Results

The results show that although MDT care is not cost effective at a £20,000 per quality-adjusted life year (QALY) threshold in the base case, there is significant uncertainty surrounding this finding as detailed in the sensitivity analyses detailed in Appendix M.

Taking into account the sensitivity of the model results in relation to changes in quality of life along with the fact that the incremental cost-effectiveness ratio (ICER) is below £30,000 per QALY in the base case, it is likely that MDT care is a cost-effective intervention, under the NICE reference case.

Refer to Section 9.4 in the full guideline for discussion of the results and limitations.

## Method of Guideline Validation

External Peer Review

Internal Peer Review

## Description of Method of Guideline Validation

### Validation Process

This guidance is subject to a 6-week public consultation and feedback as part of the quality assurance and peer review of the document. All comments received from registered stakeholders are responded to in turn and posted on the National Institute for Health and Care Excellence (NICE) Web site.

## Evidence Supporting the Recommendations

### Type of Evidence Supporting the Recommendations

Refer to "Type of Studies" in the "Description of Methods Used to Collect/Select the Evidence" field.

## Benefits/Harms of Implementing the Guideline Recommendations

### Potential Benefits

The guideline recommendations aim to improve care by maintaining functional ability and enabling people with motor neurone disease (MND) and their family members to live life as fully as possible. Early diagnosis, without delay after investigation, may be helpful as it allows for the provision of medication and aids, as well as for communication about the disease and advance care planning to be undertaken appropriately. Particular emphasis is placed on determining the best way to organise the care and management of people with MND.

See also the "Trade-off between clinical benefits and harms" sections in the full version of the guideline (see the "Availability of Companion Documents" field) for benefits of specific interventions.

### Potential Harms

- Harms of providing information and support to patients, families, and carers include the possibility that poorly prepared staff could undermine the efforts of the care team; the potential for professionals to lack knowledge and understanding was possible, given that they may not have cared for many people with motor neurone disease (MND), which worried patients.
- The evidence showed there were clinical harms for drowsiness, weakness and cramps for gabapentin.
- Three studies that compared glycopyrrolate versus placebo and 1 study of benzotropine versus placebo found both drugs to be effective for caregiver assessment of severity of drooling. However, a clinically significant number of patients discontinued the treatments due to side

effects.

- A gastrostomy can be psychologically difficult for people to cope with. The Guideline Development Group (GDG) also highlighted that on occasions whereby a person chooses to decline a gastrostomy, the healthcare professional should discuss with them the possible clinical harms and risks of a late gastrostomy or no gastrostomy: for example, low critical body mass, respiratory complications, risk of dehydration, different methods of insertion, and a higher risk of mortality and procedural complications.
- Harms may be caused by the provision of inappropriate equipment and delay in provision of equipment.

See also the "Trade-off between clinical benefits and harms" sections in the full version of the guideline (see the "Availability of Companion Documents" field) for additional detail on harms of specific interventions.

## Qualifying Statements

### Qualifying Statements

- The recommendations in this guideline represent the view of the National Institute for Health and Care Excellence (NICE), arrived at after careful consideration of the evidence available. When exercising their judgement, professionals are expected to take this guideline fully into account, alongside the individual needs, preferences and values of their patients or service users. The application of the recommendations in this guideline is not mandatory and the guideline does not override the responsibility of healthcare professionals to make decisions appropriate to the circumstances of the individual patient, in consultation with the patient and/or their carer or guardian.
- Local commissioners and/or providers have a responsibility to enable the guideline to be applied when individual health professionals and their patients or service users wish to use it. They should do so in the context of local and national priorities for funding and developing services, and in light of their duties to have due regard to the need to eliminate unlawful discrimination, to advance equality of opportunity and to reduce health inequalities. Nothing in this guideline should be interpreted in a way that would be inconsistent with compliance with those duties.

## Implementation of the Guideline

### Description of Implementation Strategy

An implementation strategy was not provided.

### Implementation Tools

Clinical Algorithm

Foreign Language Translations

Mobile Device Resources

Patient Resources

Resources

For information about availability, see the *Availability of Companion Documents* and *Patient Resources* fields below.

## Institute of Medicine (IOM) National Healthcare Quality Report Categories

### IOM Care Need

End of Life Care

Living with Illness

## IOM Domain

Effectiveness

Patient-centeredness

Timeliness

## Identifying Information and Availability

### Bibliographic Source(s)

National Clinical Guideline Centre. Motor neurone disease: assessment and management. London (UK): National Institute for Health and Care Excellence (NICE); 2016 Feb 24. 47 p. (NICE guideline; no. 42).

### Adaptation

Not applicable: The guideline was not adapted from another source.

### Date Released

2016 Feb 24

### Guideline Developer(s)

National Clinical Guideline Centre - National Government Agency [Non-U.S.]

### Source(s) of Funding

The National Institute for Health and Care Excellence (NICE) funds the National Clinical Guideline Centre (NCGC) and thus supported the development of this guideline.

### Guideline Committee

Guideline Development Group (GDG)

### Composition of Group That Authored the Guideline

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## Financial Disclosures/Conflicts of Interest

At the start of the guideline development process all Guideline Development Group (GDG) members declared interests including consultancies, fee-paid work, share-holdings, fellowships and support from the healthcare industry. At all subsequent GDG meetings, members declared new and arising conflicts of interest.

Members were either required to withdraw completely, or for part of the discussion, if their declared interest made it appropriate. The details of declared interests and the actions taken are shown in Appendix B (see the "Availability of Companion Documents" field).

## Guideline Status

This is the current release of the guideline.

This guideline updates a previous version: Centre for Clinical Practice. Motor neurone disease. The use of non-invasive ventilation in the management of motor neurone disease. London (UK): National Institute for Health and Clinical Excellence (NICE); 2010 Jul. 127 p. (Clinical guideline; no. 105). [53 references]

This guideline meets NGC's 2013 (revised) inclusion criteria.

## Guideline Availability

Available from the [National Institute for Health and Care Excellence \(NICE\) Web site](#) . Also available for download in ePub or eBook formats from the [NICE Web site](#) .

## Availability of Companion Documents

The following are available:

- Motor neurone disease: assessment and management. Full guideline. London (UK): National Institute for Health and Care Excellence; 2016 Feb. 319 p. (NICE guideline; no. 42). Available from the [National Institute for Health and Care Excellence \(NICE\) Web site](#) .
- Motor neurone disease: assessment and management. Appendices. London (UK): National Institute for Health and Care Excellence; 2016 Feb. 468 p. (NICE guideline; no. 42). Available from the [NICE Web site](#) .
- Motor neurone disease: assessment and management. Costing report. London (UK): National Institute for Health and Care Excellence; 2016 Feb. 5 p. (NICE guideline; no. 42). Available from the [NICE Web site](#) .
- Motor neurone disease: assessment and management. Baseline assessment tool. London (UK): National Institute for Health and Care Excellence; 2016 Feb. (NICE guideline; no. 42). Available from the [NICE Web site](#) .
- The guidelines manual 2012. London (UK): National Institute for Health and Care Excellence (NICE); 2012 Nov. Available from the [NICE Web site](#) .

## Patient Resources

The following is available:

- Motor neurone disease: assessment and management. Information for the public. London (UK): National Institute for Health and Care Excellence (NICE); 2016 Feb. 16 p. (NICE guideline; no. 42). Available from the [National Institute for Health and Care Excellence](#)

(NICE) Web site . Also available for download in ePub and eBook formats from the NICE Web site . Also available in Welsh from the NICE Web site .

Please note: This patient information is intended to provide health professionals with information to share with their patients to help them better understand their health and their diagnosed disorders. By providing access to this patient information, it is not the intention of NGC to provide specific medical advice for particular patients. Rather we urge patients and their representatives to review this material and then to consult with a licensed health professional for evaluation of treatment options suitable for them as well as for diagnosis and answers to their personal medical questions. This patient information has been derived and prepared from a guideline for health care professionals included on NGC by the authors or publishers of that original guideline. The patient information is not reviewed by NGC to establish whether or not it accurately reflects the original guideline's content.

## NGC Status

This NGC summary was completed by ECRI on February 4, 2011. This summary was updated by ECRI Institute on May 26, 2016.

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